Pseudo Foster-Kennedy

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Abstract

Foster-Kennedy syndrome is a condition characterized by monocular vision loss caused by direct pressure by a tumor on the optic nerve. Subsequently, due to intracranial hypertension, the contralateral optic nerve is affected, causing progressive vision loss. In the case we will discuss, we present a patient with vision loss and optic nerve atrophy secondary to idiopathic intracranial hypertension, who subsequently began to experience progressive contralateral monocular vision loss, mimicking a Foster-Kennedy syndrome. The patient ultimately required a ventriculoperitoneal shunt due to a high risk of total blindness. **(Acta Med Colomb 2024; 49. DOI: https://doi.org/10.36104/amc.2024.2998).**

Keywords: *pseudo Foster-Kennedy syndrome, idiopathic intracranial hypertension, optic nerve, visual disturbances, headache.*

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Introduction

Foster-Kennedy syndrome (FKS) is a group of signs and symptoms characterized by progressive unilateral visual acuity loss. This loss is a result of atrophy triggered by direct compression of the optic nerve, generally caused by a compressive lesion in the anterior fossa, associated with contralateral papilledema secondary to increased intracranial pressure (1). In addition, patients may have headaches, anosmia and/or tinnitus (2). There is an alternative classification which includes the described signs and symptoms in the absence of a compressive lesion; this specific type is known as "Pseudo Foster-Kennedy" (2).

The main causes of Pseudo Foster-Kennedy syndrome include optic neuritis, trauma, vitamin B12 deficiency (3), neurosyphilis and idiopathic intracranial hypertension (4). Below, we will present the case of a 46-year-old man who developed progressive visual acuity loss and anosmia due to idiopathic intracranial hypertension.

Case presentation

A 46-year-old male patient with no significant past medical history had experienced progressive visual acuity loss over the previous 10 months, beginning in his left eye and subsequently affecting his right eye, coupled with tinnitus and anosmia. On physical exam, he had no vital sign abnormalities. He had a Marcus Gunn pupil in his left eye, with shape vision at one meter and finger counting acuity at less than 20 cm distance; he had a hyporeactive pupil and papilledema in the right eye, with preserved visual acuity (20/20). The fundal exam showed left optic nerve atrophy and papilledema in the right eye.

He underwent magnetic resonance imaging of the brain and orbits showing no compressive lesions, evidence of an empty sella and dilation of the optic nerve sheath (Figure 1). A magnetic resonance angiography of the brain ruled out venous sinus thrombosis and the Humphrey visual field test showed the patient's visual defect (Figure 2). Complementary laboratory tests were within normal limits. In addition, a lumbar puncture was performed with an opening pressure of 46 cm H_2O, and no pathological findings in the cerebrospinal fluid (Table 1).

In light of the patient's signs and symptoms and the procedures performed, particularly the elevated lumbar puncture opening pressure with no trigger in the complementary tests, a primary diagnosis of idiopathic intracranial hypertension was made. Consequently, treatment was started with acetazolamide 250 mg/12 hours with no symptom improvement. Subsequently, due to the high risk of complete vision loss, a ventriculoperitoneal shunt was ordered, in conjunction with the neurosurgery department, which was placed without complications. Ultimately, the patient had no progression of the contralateral visual defect.

Discussion

Foster-Kennedy syndrome, as a set of signs and symptoms, has the cardinal symptom of progressive unilateral loss of visual acuity caused by atrophy secondary to direct compression of the optic nerve, produced by a compressive mass in the anterior fossa and associated with contralateral papilledema secondary to increased intracranial pressure (1). In addition, 84% of patients may have headaches, which may be described as migraines (52%) or tension headaches (22%); as well as tinnitus (52%) or anosmia (2–5). In the case

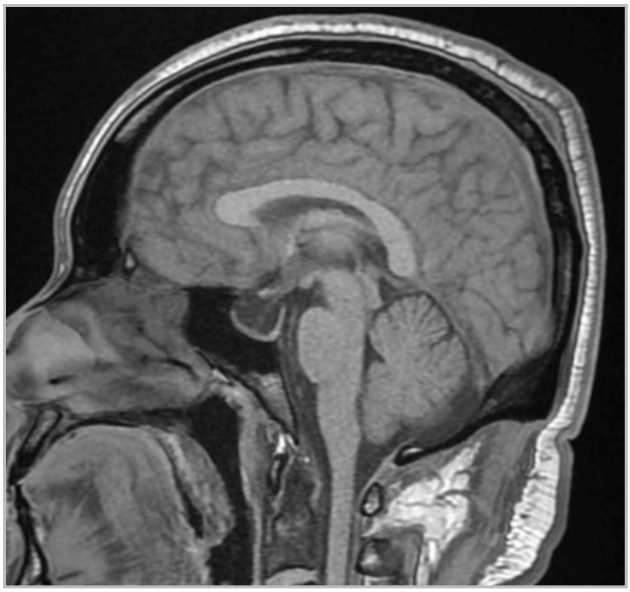

Figure 1. *Magnetic resonance imaging of the brain with T1 sequences in the sagittal plane showing an empty sella turcica..*

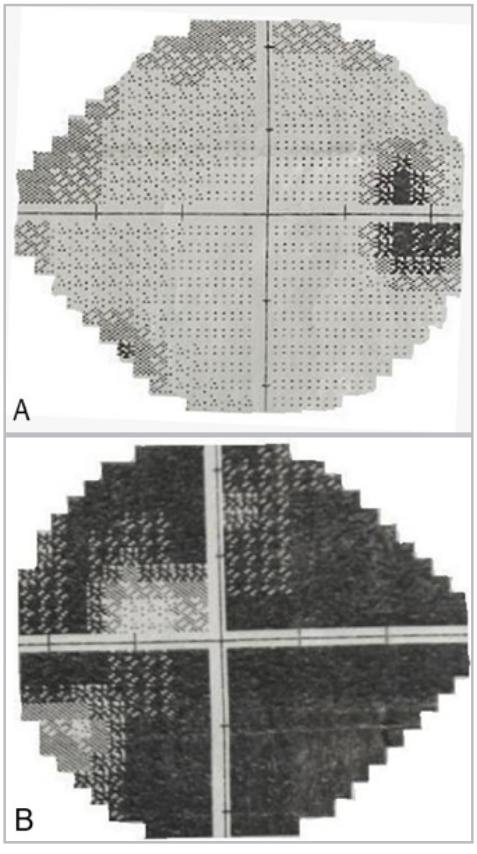

Figure 2. A. *Right eye*. **B**. *Left eye*. *The Humphrey visual fields show visual field loss in the left eye with a minimum of 0 decibels (dB) in the darkest areas and a maximum of 24 dB in the upper right quadrant (lighter areas). In addition, there is a minimum of 5 dB and a maximum of 30 dB in the right eye, within normal limits.*

Table 1. Laboratory tests performed on the patient and their reference values.

Laboratory tests	Patient's results	Reference values
Hemoglobin (g/dL)	16.12	14 - 18
Hematocrit (%)	46	42 - 50
Mean corpuscular volume (fL)	93.8	80 - 98
Mean corpuscular hemoglobin (pg/cell)	32.2	28 - 32
Platelet count (10^3/uL)	282	150 - 500
Total leukocytes (cell/microliter)	8,020	5 - 10
Neutrophils (cell/microliter)	4,210	1.4 - 6.5
Lymphocytes (cell/microliter)	3,100	1.2 - 3.4
Serum creatinine (mg/dL)	1.09	0.7 - 1.3
C-reactive protein (mg/dL)	0.4	<0.8
Serum glucose (mg/dL)	97	70 - 99
PTT (seconds)	25.2	25 - 35
PT (seconds)	10.3	11 - 13
TSH (uIU/mL)	1.54	0.5 - 4
Vitamin B12 (pg/mL)	432	232-1245
HIV ELISA	Nonreactive	Nonreactive
VDRL	Nonreactive	Nonreactive
Antinuclear antibodies	Nonreactive	Nonreactive
Anti-Ro antibodies	Nonreactive	Nonreactive
Anti-La/SSB antibodies	Nonreactive	Nonreactive
Anti-Sm antibodies	Nonreactive	Nonreactive
Anti-RNP antibodies	Nonreactive	Nonreactive
CSF cytochemical testing	Colorless - transparent Leukocytes: 0 - erythrocytes: 0 Glucose: 61 mg/dL Protein: 35.9 mg/dL	
CSF VDRL	Nonreactive	Nonreactive
CSF Gram	Negative	Negative
CSF adenosine deaminase	0.2 U/L	0 to 32 U/L
CSF bacilloscopy	Negative	Negative
CSF cryptococcal antigen	Nonreactive	Nonreactive

PTT: partial thromboplastin time; PT: prothrombin time; INR: international normalized ratio; TSH: thyroid stimulating hormone; ADA: adenosine deaminase; CSF: cerebrospinal fluid; HIV: human immunodeficiency virus; VDRL: Venereal Disease Research Laboratory, a non-treponemal test for syphilis; g: grams; mg: milligrams; pg: picograms; dL: deciliter; mL: milliliter; fL: femtoliter; cell: cells.

described, the patient initially presented with chronic vision problems in his left eye and acute deterioration of the visual fields in his right eye. The physical exam showed right unilateral papilledema with associated left optic nerve atrophy. The Humphrey visual fields showed visual field loss in the left eye with a normal visual field in the right. The patient also reported having anosmia.

The FKS classification also includes Pseudo Foster-Kennedy, which is defined as a condition in which the patient manifests all the characteristic signs and symptoms, with no compressive mass on neuroimaging (2). This patient had progressive vision loss with left optic nerve atrophy and subsequent involvement of the right optic nerve, coupled with anosmia. It is notable that there was no headache in this

case, in the context of a patient with a brain MRI suggestive of long-term intracranial hypertension, with no compressive mass on neuroimaging. In addition, infectious, autoimmune and hematological causes were ruled out as etiologies of the intracranial hypertension (Table 1).

The normal neuroimages found in cases of idiopathic intracranial hypertension (IIH) include several characteristic ones like an empty sella turcica; flattening of the posterior globe and protrusion of the optic nerve (with 57% sensitivity, but the highest specificity of all the radiological characteristics, at 97%), optic nerve sheath dilation with 51% sensitivity and 83% specificity; and distal transverse sinus stenosis, with 78% sensitivity and indeterminate specificity. All these findings suggest long-standing intracranial hypertension (6). Furthermore, if at least three of these characteristics are present, they reach an almost 100% specificity, maintaining an approximate sensitivity of 64% (7). In the specific case of our patient, Figure 1 shows an empty sella turcica and enhancement of the optic nerve sheath, predominantly on the left.

Magnetic resonance imaging of the brain should be performed to look for lesions that could be causing intracranial hypertension. Possible etiologies include mass-effect lesions, hydrocephalus, venous sinus thrombosis, arteriovenous fistulas, bilateral jugular vein obstruction and subarachnoid hemorrhages (5). Studies were done on our patient confirming the absence of these lesions.

A high opening pressure was recorded on lumbar puncture, and complementary cerebrospinal fluid testing was done (Table 1). The results showed cerebrospinal fluid with no pathological characteristics. Magnetic resonance imaging of the brain also ruled out bleeds, venous sinus problems and expansive masses, thus establishing the diagnosis of idiopathic intracranial hypertension, meeting the modified Dandy criteria (Table 2). It should also be noted that the Friedman criteria can be used for diagnosis when the patient does not have optic nerve involvement (8, 9).

Idiopathic intracranial hypertension has shown a seven times greater risk of associated vision defects compared to the general population, with an OR of 8 (95% CI 3.7–17.1) (10). As far as treatment for IIH, there are various medical and surgical lines of therapy.

The first line of therapy includes medications like acetazolamide, which inhibits carbonic anhydrase in the choroid plexus, affecting CSF secretion (6). Other options include corticosteroids, which are not recommended for long treatment courses. Topiramate contributes to headache treatment, weight control and carbonic anhydrase reduction (6).

As far as surgical treatment, the most common indication for surgery is persistent deterioration despite optimal medical treatment. The options include cerebrospinal fluid shunting and optic nerve sheath fenestration. Although the latter option is recommended when headaches are very severe, the choice is left up to the availability and experience of the local team (6).

Table 2. *Modified Dandy criteria for diagnosing idiopathic intracranial hypertension (9).*

1.	Signs and symptoms of intracraneal hypertension: headache, changes in visual acuity and papilledema.
2.	No focal neurological signs, except sixth cranial nerve paresis.
3.	Elevated cerebrospinal fluid opening pressure on lumbar puncture (>25 cm H_2O).
4.	Normal or small symmetrical architecture of the cerebral ventricles on neuroimaging, with no masses or venous system obstruction.
5.	No other etiology of increased intracranial pressure.

The only measure considered to have an impact on recovering lost vision, when diagnosed early, is cerebrospinal fluid shunting (11). In our patient's case, this was the intervention performed.

Conclusion

Foster-Kennedy is a syndrome characterized by damage to the optic nerves due to increased intracranial pressure, generally secondary to a tumor mass in the central nervous system. However, this syndrome may also occur from any cause of persistent intracranial hypertension. Treatment of this condition should be aimed at controlling headaches and preserving vision. In this case, a Pseudo Foster-Kennedy syndrome due to IIH was treated with early ventriculoperitoneal shunting, with the treatment goal of preserving vision.

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